Monday, March 4, 1991
2:00PM–3:30PM, Room 260, West Concourse
Natural and Unnatural History of Pediatric Heart Disease

2:00

EXTENDED LONG TERM FOLLOW-UP STUDY OF CORONARY ARTERIAL LESIONS IN KAWASAKI DISEASE

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For the purpose of studying the course of coronary arterial lesions in Kawasaki disease, the time of appearance of stenotic lesions on coronary arteriography (CAG) was analyzed based on 193 patients who underwent more than three CAGs (3.8 ± 1.1 times; mean ± SD) and 7 deceased cases who died before their third CAG. Their period of follow-up ranged from 6 months to 15 years (96.2 ± 43.7 months). Stenotic lesions were classified into 3 groups, such as occlusion (100% obstruction), segmental stenosis (recanalization) and localized stenosis of greater than 90% (≧90% LS). Occlusion of the left coronary artery (LCA) was seen in 28 patients (14%), while the right coronary artery (RCA) was occluded in 36 patients (18%). Most of the occlusion appeared within 2 years of the onset of the disease. The incidence of segmental stenosis in RCA (22%) was significantly higher than that in LCA (3%)($p<0.01$). All the segmental stenosis appeared within 7 years of the onset of the disease. ≧90% LS was seen significantly more frequently in LCA (13%) than in RCA (3%). The CAG obtained more than 8 years after the onset of the disease showed the highest incidences of 90% LS in LCA.

Conclusions: Coronary arterial lesions on RCA and that on LCA showed the different course. Occlusion of RCA tend to recanalize and LS of LCA tend to progress gradually even several years after the onset of the disease. Aneurysm of LCA, therefore, should be followed carefully for a long period of time by CAG.

2:15

FACTORS ASSOCIATED WITH IMPROVED SURVIVAL AFTER MODIFIED FONTAN OPERATIONS

John E. Mayer,Jr, Nancy D. Bridges, James E. Lock, Richard A. Jonas, and Aldo R. Castaneda, Children's Hospital, Boston, MA.

Review of current results (7/88-6/90) of modified Fontan operations showed a remarkable reduction in mortality risk to 3.2% (4/124, group II) compared with the prior two years 16% risk (14/87, group I). To understand the factors associated with this improvement, comparison was made for number of risk factors (RF) (anomalies of venous drainage, age<4 yrs, PA pressure >15mmHg, pulmonary vascular resistance >2 Wood units (indexed), and PA distortion), and for the effects of technical modifications including cavopulmonary connection (CPCx), fenestration of interatrial baffle (Fen), use of an intra-atrial cavo-caval (C-C) tubular conduit to segregate systemic venous return, and use of bidirectional cavopulmonary Glenn (G) shunt as a takedown procedure (TD) for a failing Fontan instead of a systemic to pulmonary shunt (S). (Table numbers refer to death/total)

Grp	Total	noRF	1RF	>1RF	CPCx	C-C	Fen	TDS	TDG
I	14/87	1/30	5/43	8/14	4/12	0/2	0/0	2/2	0/0
II	4/124	0/32	1/57	3/32	2/112	3/89	2/51	0/1	1/6

We conclude that more stringent patient selection did not account for the improved results since a higher % in the later group had 2 or more RF, but management modifications and a more aggressive stance toward taking down a failing Fontan to a bidirectional Glenn shunt were associated with the improved survival results.

2:30

FONTAN PROCEDURE FOLLOWING BIDIRECTIONAL GLENN SHUNT FOR HIGH RISK PATIENTS

Richard A. Jonas, Nancy D. Bridges, Frank L. Hanley, John E. Mayer, Aldo R. Castaneda
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We recently reported the low mortality and morbidity of intermediate procedures (bidirectional Glenn shunt (BDG), fenestrated Fontan (FF)) in patients with multiple risk factors for a Fontan procedure. However, there are few data regarding the ability of a BDG to prepare a "high risk" patient for subsequent Fontan procedure. Between June, 1988 and May, 1990, 60 patients had a BDG at the Children's Hospital, Boston. Between May 1989 and July 1990, 13 of them underwent Fontan procedures (9 FF, 4 non-fenestrated), mean 13 months following BDG. 8 of the 13 patients had either hypoplastic left heart syndrome or double outlet right ventricle with mitral atresia. Mean age of patients at the time of BDG was 35 ± 18 months. All patients had had at least one previous palliative procedure before BDG. Risk factors before the BDG included pulmonary artery hypoplasia or distortion in 7, ventricular dysfunction in 5, pulmonary vascular resistance (PVR) elevated in 4. Mean PVR for the group varied little from 2.04 ± 0.61 before BDG to 1.98 ± 1.21 mean 11.5 months following BDG ($p = 0.87$). Mean end diastolic pressure was 9.2 ± 3.4 mm before BDG and was 8.4 ± 2.8 mean 11.5 months after BDG ($p = 0.5$). One patient had dramatic improvement in atrioventricular valve regurgitation. There was one early death after Fontan procedure. One patient had takedown to BDG because of low cardiac output. Median hospital stay was 23 days (mean 26 days). 4 of 12 survivors drained pleural effusions for more than one week. There have been no late deaths. One patient continues to have choreoathetosis which was present after BDG.

In conclusion, while the BDG will represent final palliation for some very high risk patients, selected patients may have subsequent completion of the Fontan operation with acceptable risk.

2:45

OUTCOME FOLLOWING BIDIRECTIONAL CAVO-PULMONARY ANASTOMOSIS PRIOR TO MODIFIED FONTAN PROCEDURE

Jeanne Marie Balfa, Jack Rychik, Scott D. Gullquist, Gerald Barber, Marshall L. Jacobs, William I. Norwood, John D. Murphy, *The Children's Hospital of Philadelphia, Philadelphia, PA, USA.*

A bidirectional cavo-pulmonary anastomosis or "Hemi-Fontan"(HF) has been used at our institution as an intermediate step prior to modified Fontan procedure(FP) in order to acheive earlier reduction of volume load on the ventricle and to minimize the impact of the geometric changes which may occur in response to this volume diminution on the outcome of eventual FP. Between May 10, 1989 and July 31, 1990, 114 consecutive pts underwent HF in anticipation of eventual FP. The operation consisted of anastomosis of the right superior vena cava/right atrial junction to the right pulmonary artery with homograft augmentation of the pulmonary arteries and patch closure of the superior vena caval entrance into the right atrium. Anatomic diagnoses in the patient population included hypoplastic left heart syndrome(67), double outlet right ventricle(10), transposition of the great arteries (S,L,L)(10), tricuspid atresia(8), pulmonary atresia/intact ventricular septum(5) and other(14). Age range at HF was 1.5 to 88 mos with a median age of 11.4 mos. The median hospital stay following surgery was 7d (range 4 to 95d). Before HF, 110 of the pts underwent 119 surgical procedures including Norwood Stage I reconstruction(82), AO to PA shunt(20), PA banding(8) and balloon atrial septotomy(5). Cardiac catheterizations were performed in 108 of the pts prior to HF. Subsequently, 41 pts underwent cardiac catheterization in anticipation of FP. The hemodynamic data are presented in the table.

	AO saturation	PA pressure	EDP
Pre HF:	78% ±6	18 ±10	10 ±3
Post HF:	82% ±7	13 ±5	10 ±4
	[p<0.01]	[p<0.01]	[p=NS]

There were 13 deaths during the study period, with an actuarial survival of 89% at 6 mos and 84% at 12 mos. Of the pts that died, 7 had coexisting anatomic or hemodynamic problems, notably, residual coarctation(2), ventricular dysfunction(2), aneurysmal dilation of the proximal arch(1), coronary thrombosis(1), and severe pulmonary hypertension(1).

Conclusion: Hemi-Fontan has low morbidity and mortality and yields favorable hemodynamics and oxygenation. We speculate that this interim step may improve overall morbidity and mortality of subsequent Fontan operation.